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Brain infarction following elective laparoscopic cholecystectomy in a patient with sickle cell disease and previously undetected Moyamoya syndrome☆

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ABSTRACT

Objective: We report a 7 year-old boy, known case of sickle cell disease, who underwent laparoscopic cholecystectomy for gallstones. He sustained brain insult due to undiagnosed MoyaMoya syndrome. The purpose of this report is to familiarize the healthcare community with similar events and discuss possible plans/recommendations for future similar cases.

Method: Case report and review of the English-language literatures (using PubMed, Ovid, and Proquest databases).

Results: Case of complicated MoyaMoya syndrome sustained brain insult postoperatively in a 7 year-old boy, with sickle cell disease and gallstones.

Conclusion: Early onset of MoyaMoya syndrome related stroke postoperatively after laparoscopic cholecystectomy is difficult to be assessed immediately. Acute stroke is an infrequent disease of pediatric age group patients. MoyaMoya is a rare cerebrovascular disease of unknown etiology. Our patient is 7 year-old male child who presented immediately postoperatively with abnormal movement of right upper and lower limbs and right upper limb weakness, and no history of MoyaMoya syndrome, which is rare, but important cause of stroke in children. Cerebral revascularization surgery leads to favorable outcome. The present case highlights the importance of considering "MoyaMoya syndrome" in sickle cell disease patients who are going for surgery irrespective of their age group.

1. Introduction

Sickle cell disease (SCD) in certain parts of the world is the most common genetic disorder. It is believed that 1.7% of the population of the Eastern Province of Saudi Arabia is affected and they are at an increased risk of developing pigmented gallstones, this risk increases with age [1]. At our institution average 2500 sickle cell disease patients less than 15 years old are annually visiting our hematology clinic. Laparoscopic Cholecystectomy for both symptomatic and asymptomatic gallstones is commonly performed in patients with this disease, and it is the most frequently performed surgical procedure in this population. The occurrence of serious sickle cell events such as vaso-occlusive pain episodes and acute chest syndrome (ACS) are the usually post-surgical complications that could be encountered in those population. Brain infarction following laparoscopic cholecystectomy in sickle cell disease is a rare complication and difficult to anticipate, in this case patient found to have postoperative grade 1 MoyaMoya syndrome (MMS) which is at high risk of brain stroke. Children with sickle cell anemia are at significantly increased risk of cerebrovascular accidents. This was first described in 1923 by Sydenstriker, who reported convulsions and hemiplegia in a 5-year-old boy and left hemiparesis in a 3-year-old baby [2]. This rare complication i.e Brain infarction after laparoscopic cholecystectomy forced us to investigate more through CT scan, MRI, MRA and cerebral angiography, which led us to the diagnosis of MMS. The disease commonly presents with strokes in childhood, while in adults subarachnoid and intracranial hemorrhage is the prevailing presentation [3]. The purpose of this report is to update our experience and to continue supporting the safety of laparoscopic cholecystectomy in sickle cell patients with gallstones.

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Table 1
Describe the labs results and hemoglobin electrophoresis. The values are demonstrated as a preoperatively and postoperatively.

TCDI	AT Diagnosis 4 months	PRE- hydroxyurea 2015	POST- Hydroxyurea 2017	AT date of surgery 05/2017	Pre- exchange TX	Post- exchange TX
		normal	normal			
Hb mg/dl	6.9	7.0	9.0	8.6	9.3	11.0
Hb A%	0	0	0	0	20.6	62.4
Hb S%	65	88.7	74.9	73.6	59.8	27.4
Hb A2%	1.6	2.8	2.9	3.0	2.8	2.7
Hb F %	33.2	8.5	22.2	23.4	16.8	7.5

2. Case presentation

A 7 year-old boy diagnosed to have sickle cell disease by neonatal screening and since infancy he was on regular follow up in pediatric hematology clinic. He was receiving comprehensive care and management. Later, he was diagnosed to have Hypothyroidism and bronchial asthma. Patient is on regular medication for SCD with good compliance and had some difficult course during infancy in the form of dactylitis and recurrent admission with multiple blood transfusions for which he was started on Hydroxyurea. Last blood transfusion was 18 months back. Later on, he developed gall stones with recurrent episodes of upper abdominal pain. He was admitted electively on May 2017 for Laparoscopic Cholecystectomy. his Initial haemoglobin was 8.6 gm/dl and Hb S was 73.6% at the time of admission but after packed red blood cell transfusion, Hb became 10.8 gm/dl and Hb S 59.8% see [Table 1]. Laparoscopic cholecystectomy done for him one day after the admission. It was uneventful operation, anesthesia with 90 min total time, and smooth recovery. Patient shifted to general ward with advice of intravenous fluids and analgesia. Five hours after surgery, he developed abnormal movement of Right lower limb with weakness of Right upper limb and drooling of saliva. CBC & U&E samples showed normal results. **CT scan brain** done immediately [Fig. 1], and patient shifted immediately to intensive care unit where he underwent blood exchange transfusion next day.

Next day, **MRI brain** [Fig. 2] and **Cerebral Angiography were done** [Fig. 3].

All these findings are most consistent with Suzuki stage 1 Moyamoya syndrome. The patient stayed in the hospital for 2 weeks after cholecystectomy to start physiotherapy, speech therapy & occupational therapy. He improved power in his limbs, and started to talk gradually. Patient was discharged with a right sided hemiparesis.

3. Discussion

Gallstones are a frequent complication in children with hemoglobinopathies because of the recurrent episodes of hemolysis leading to an increase in bilirubin excretion and pigment gallstones formation. The incidence of gallstones in children with SCD increase with age, 15% under 10 years of age, 22% between 10 and 14 years of age, and 36% between 15 and 18 years of age, with a reported prevalence of 50% by the age of 22 [4]. Laparoscopic Cholecystectomy is the gold standard treatment for both symptomatic and asymptomatic gallstones. Perioperative morbidity and mortality rates higher than those noted in the general population are expected in sickle cell disease patients and it is related to increase perioperative red cell sickling. Moyamoya disease (MMD) was first described in Japan by Takeuchi and Shimizu in 1963. MMD is a chronic, progressive occlusion of the circle of Willis arteries that leads to the development of characteristic collateral vessels seen on imaging, particularly cerebral angiography as “puff of smoke”. The process of narrowing of cerebral vessels seems to be a reaction of brain blood vessels to a wide variety of external stimuli, injuries, or genetic defects. Conditions such as sickle cell anemia, neurofibromatosis-1, Down's syndrome, congenital heart defects, antiphospholipid syndrome, renal artery stenosis, and thyroiditis have been found to be associated with MMD in the literatures [5]. When there is known underlying pathology, the condition is correctly referred to as MoyaMoya

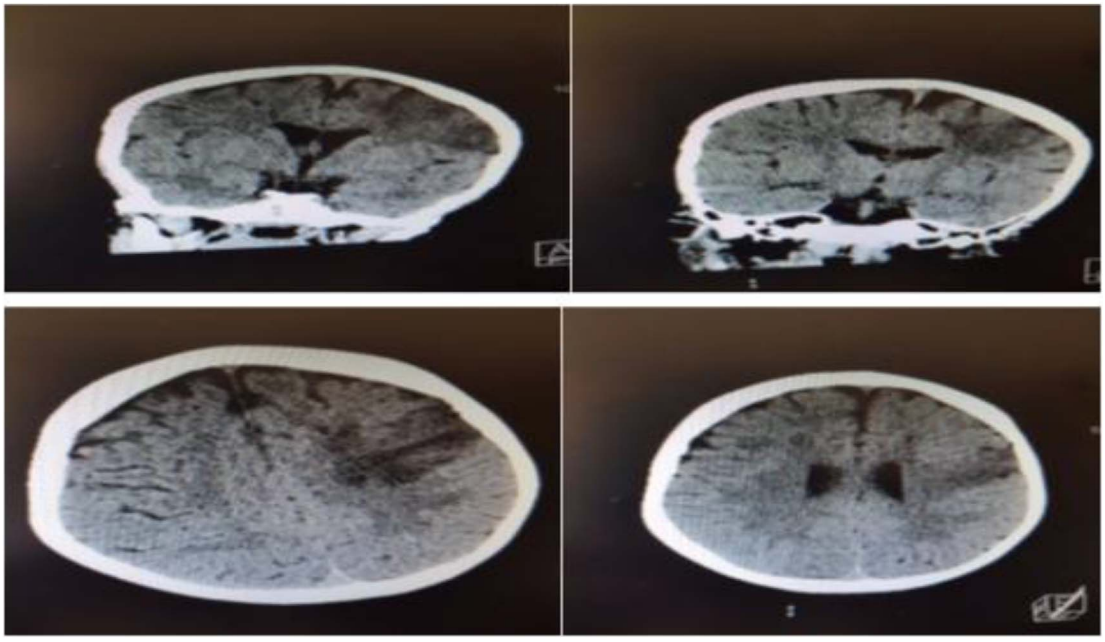

Fig. 1. CT scan brain: showed hypo-density area noted at the left frontal lobe involving cortex and white matter, representing acute infarction.